

# Discoid Lupus Erythematosus Presenting as Upper Eyelid Edema and Erythema

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Received: 15 Mar. 2016; Accepted: 14 Oct. 2016

**Abstract:** Discoid Lupus Erythematosus (DLE) is an autoimmune disorder that usually occurs on sun exposed areas of head and neck. Infrequently it could be presented by palpebral involvement and rarely unilateral upper eye lid edema and erythema have been reported as the sole manifestation of DLE. We describe a 38-year-old woman with chronic left upper eye lid edema and erythema from one year ago which was induced by steroid injection for left eyebrow alopecia. Histopathologic and direct immunofluorescent studies were made on palpebral skin tissue and confirmed DLE diagnosis. Antinuclear antibody (ANA) titer was 1/160 with speckled pattern. She was treated by oral hydroxychloroquine (400 mg daily) with moderate improvement after three months. We should think about DLE in cases with chronic upper eye lid edema and erythema. The aim of this case report is to emphasize that ophthalmologist and dermatologists should be aware of different presentations of DLE in the periorbital area to prevent misdiagnosis.

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*Acta Med Iran* 2017;55(7):474-476.

**Keywords:** Discoid lupus erythematosus; Eyelid; Edema; Erythema

## Introduction

DLE is a chronic autoimmune disorder which typically presents by circumscribed, scaly, erythematous lesions which involve sun exposed areas especially face, scalp and ears (1). DLE lesions on the eye lids were first described in 1932 (2). Gupta *et al.*, have reported palpebral lesions in only 6% of lupus cases, often associated with other cutaneous abnormalities (1) but eyelid involvement as the sole presentation of DLE is exceptionally rare. DLE on the eyelids could manifest as blepharitis, lid plaque, lid scarring, symblepharon, madarosis and periorbital edema (3,4,5). This case is reported to provide more information regarding unusual presentations of DLE.

## Case Report

We present herein a 38-year-old farmer woman with one-year history of left upper eyelid edema which was superimposed by erythema after six months. Our case mentioned that she had hair loss on the lateral side of her left eyebrow two months prior the beginning of edema. Alopecic patch was treated by intralesional

triamcinolone injection one week before upper eyelid edema, and no other trauma was noted. Eyebrow alopecia was improved three weeks after steroid injection, but eye lid edema continued and became erythematous after six months. Edema was worse in the morning and never resolved during the last year. She complained of eyelid pruritus and photophobia without any pain on the involved side.

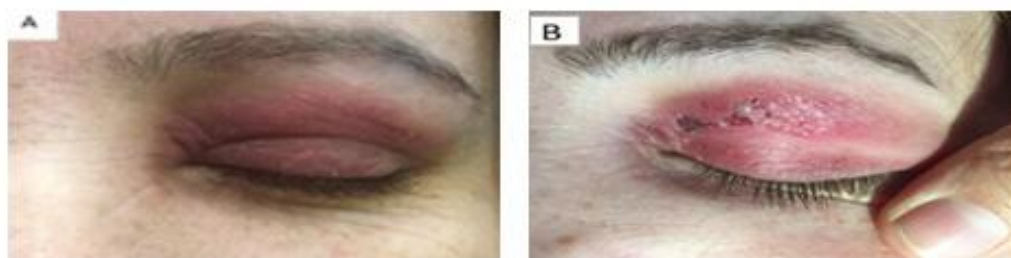
In past medical history, she had a neurosurgical procedure for a congenital abnormality in thoracic vertebrae four years ago. In drug history, she had steroid injection in the eyebrow and oral antibiotic therapy for eyelid erythema.

In physical examination, there were erythema and edema on the left upper eyelid with fine scaling in the first visit, but gradually significant scales and crust appeared especially two weeks after skin biopsy (Figure 1). She had Raynaud's phenomenon without cuticular telangiectasia.

In ophthalmologic consult regarding photophobia, only blepharitis and dry eye were detected. There was no lower lid involvement, madarosis, photosensitivity, arthralgia, arthritis or scarring alopecia

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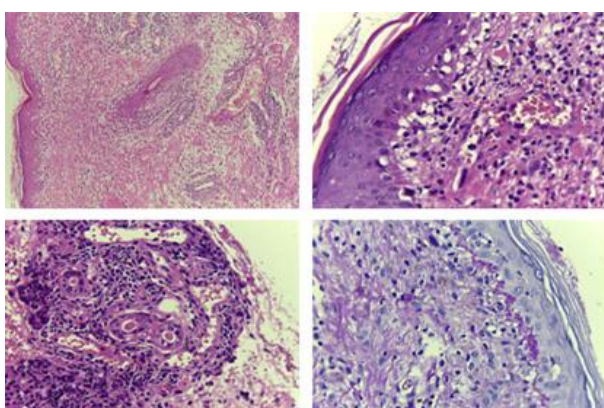
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**Figure 1.** A) Unilateral upper eyelid edema and erythema before skin biopsy. B) Some scales and crust were seen 2 weeks after skin biopsy

Skin biopsy was made with differential diagnosis of pseudolymphoma, DLE, contact dermatitis, sarcoidosis and cutaneous metastasis.

Histopathologic changes were epidermal atrophy, mild focal parakeratosis, marked basal vacuolar damage, focal basement membrane thickening, capillary dilatation, superficial and deep perivascular lymphoplasmic cells infiltrate with periadnexal and perineural involvement (Figure 2).

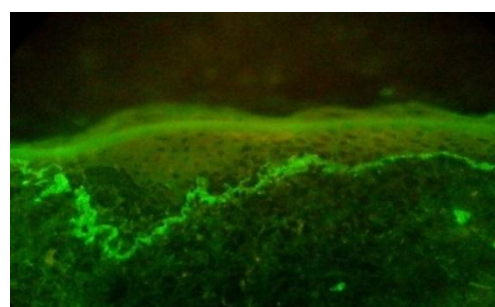


**Figure 2.** A) Low power view showing interface dermatitis with superficial and deep perivascular infiltrate of lymphoplasmic cells with follicular involvement. B) Close up view of the interface changes and telangiectasia. C) Perieccrine inflammation. [Hematoxylin-eosin, original magnification: (A)×10, (B)×40, (C)×40]. D) Focal basement membrane thickening was seen. [Periodic acid Schiff staining, original magnification: ×40]

Alcian blue staining was negative for mucin deposition. In immunohistochemistry study the infiltrate showed mixed B and T cells nature. The direct immunofluorescent study showed granular antibody deposits along dermoepidermal junction included mostly immunoglobulin G, immunoglobulin M, C3, C4 and few immunoglobulin A (positive lupus band) (Figure 3). So the diagnosis of chronic cutaneous lupus erythematosus

was confirmed. Antinuclear antibody (ANA) titer was

1/160 with the speckled pattern, but anti-double stranded DNA antibody titer was within normal limit. Mild anemia due to minor thalassemia was detected. Leukocyte, platelet, reticulocyte count, kidney function test, erythrocyte sedimentation rate, C-reactive protein, complement levels and urine analysis were normal. Orbital imaging and chest x-ray had no remarkable finding.



**Figure 3.** Immunofluorescent staining of left upper eyelid section demonstrated coarse granular thick band of immunoglobulin M along the dermoepidermal junction (lupus band). (×200, direct immunofluorescence)

## Discussion

DLE develops as a localized lesion commonly above the neck on photoexposed areas (6). This case was interesting to us because upper eyelid edema and erythema as the sole manifestation of DLE are exceptionally rare. Periocular DLE presents with a wide clinical variation and mimics blepharoconjunctivitis, cellulitis, allergic contact dermatitis, psoriasis, atopic dermatitis, ocular tumors or sebaceous carcinoma. Therefore high index of suspicion is mandatory to make the proper diagnosis (1). The eyelid DLE may have pruritus and gets aggravated with sun exposure and trauma (7). In our case, trauma due to steroid injection

could be the initiating factor to induce DLE and skin biopsy aggravated scaling of the lesion. We did not find persistent unilateral upper eyelid edema as a DLE presentation in prior studies and only a 47-year-old woman with intermittent left upper lid swelling has been reported (1). In the previous case reports periorbital swelling, itching, erythema, lower eye lid edema associated with malar edema, thickening of upper and lower eyelids, madarosis, eyelid dyspigmentation and symblepharon have been detected (1,2,4,5,8,9). In a systematic review by Arrico *et al.*, in 2015 blepharitis was the most common manifestation of ocular lupus, and the lower eyelid was the most common site of involvement. Isolated upper lid involvement was reported only in one case. Extra ocular involvement was detected in 93% of cases (10). Peggy *et al.*, reported dry eye in 7% of cases with ocular chronic cutaneous lupus erythematosus (3), similarly to our patient.

Arrico *et al.*, reported increasing in ANA titer in 21.1% of cases mostly with speckled pattern in agreement with our patient (10). The median diagnostic delay was 38 months in a case series (1), fortunately, in our case, this interval was shorter (12 months) without any scarring and madarosis. So proper differential diagnoses in cases with chronic and refractory blepharitis or eyelid edema prevent the misdiagnosis and delay in the treatment. Therefore ophthalmologists and dermatologists should be aware of the ocular manifestations of DLE.

Oral hydroxychloroquine, topical corticosteroids, intralesional corticosteroids have been effective in prior studies (1,4,7,10). Therefore hydroxychloroquine (400 mg daily) was started for her and protection against sun exposure was advised. She had moderate improvement three months later.

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